Clinical Medicine

Causalgic Form of Postphlebitic Syndrome

A Variety of Reflex Sympathetic Dystrophy Caused by Acute Deep Thrombophlebitis

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The causalgic form of the postphlebitic syndrome or reflex sympathetic dystrophy resulting from acute deep thrombophlebitis is a relatively uncommon and, unfortunately, frequently unrecognized form of the postphlebitic syndrome. The usual signs of venous insufficiency are minimal, but severe burning pain is characteristic, usually increased by dependency. The diagnosis is confirmed by phlebography and the response to a lumbar sympathetic block. A lumbar sympathectomy produces permanent pain relief.

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In 1945, a number of patients were admitted from medical facilities overseas to the DeWitt General Hospital, a United States Army vascular surgical center, for the treatment of the consequences of acute deep thrombophlebitis of the lower extremities. Five of these patients showed little or none of the usual signs of the postphlebitic syndrome—edema or venous stasis changes—but they all complained of unusual lower extremity pain. The notable subjective pain with a paucity of objective findings led to a suspicion by the surgical staff of some degree of malingering. When a phlebography was done, the existence of the postphlebitic syndrome was established in every case. It was observed that all of these patients manifested high vasomotor tone, usually greater in the affected lower extremity.

Because of the resemblance to causalgia, the therapeutic effect of a lumbar sympathetic block using procaine hydrochloride was tested. In each instance, striking relief of pain was reported that lasted several hours after the vasomotor effect of the block had disappeared. Two more blocks produced more striking relief in one case so that the pain seemed permanently relieved. The response to repeated blocks was not lasting in the other four cases, but a lumbar sympathectomy with removal of ganglia 1, 2, and 3 provided permanent pain relief in each case.

Since the end of World War II, I have seen several hundred patients with the postphlebitic syndrome. Among this rather large number, 14 have had the causalgic form first noted in the Army hospital in 1945. While this form of a postphlebitic state is relatively uncommon, it is not so rare as to be unrecognized. Nevertheless, only one of seven excellent monographs in vascular surgery,[1-7] that by Bergan and Yao,[6] mentions the syndrome. As recently as September 1987, a review of the reflex sympathetic dystrophy syndrome by Smith and Campbell listed 22 precipitating events or conditions associated with reflex sympathetic dystrophy but did not include acute deep thrombophlebitis or the postphlebitic syndrome.[8] Because the causalgic form of the postphlebitic syndrome seems too little known, this review is presented.

Clinical Features

- Usually a patient has a history of acute deep thrombophlebitis of a lower extremity in which treatment was delayed or inadequate.
- The chief complaint is severe burning pain in the extremity below the knee. The usual symptoms of venous insufficiency are slight or absent.
- An objective examination reveals high vasomotor tone in both lower extremities, greater on the affected side. Calf tenderness is frequent without the other signs attributed to acute deep thrombophlebitis. There may be some edema, but the skin changes of chronic venous insufficiency are absent.
- A plain x-ray film of the bones in the extremity will show diffuse atrophy in about half the cases (6 of 14). The spotty atrophy of Sudeck's sympathetic dystrophy is not encountered.
- A lumbar sympathetic block with a local anesthetic produces striking and sustained relief of pain lasting several hours longer than the thermal effect of the block. In some cases, a second or a third block may produce a permanent relief of pain. Most patients report a shorter duration of analgesia with repeated blocks but obtain permanent relief from a surgical sympathectomy with the removal of lumbar ganglia 1, 2, and 3. After the sympathectomy, there may be a slight increase in the manifestations of venous insufficiency that respond to the usual program of elastic support, avoiding excessive static dependency, nocturnal foot elevation, and appropriate exercise.

The following cases are characteristic:

Reports of Cases

Case 1

The patient, a 28-year-old woman, was delivered of her second baby without obvious complications and was dis-

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charged from hospital on her fourth postpartum day. No untoward symptoms were noted during her hospital stay, but the nursing records showed a labile tachycardia from 72 to 110 per minute each day on the second and third day after delivery. On the seventh postpartum day, because of pain in her lower extremity, she presented at a hospital emergency department where a painfully swollen left lower leg was reported with much calf tenderness and a positive Homans' sign. A phlebogram showed thrombosis of all of the venous channels below the knee with extension into the distal femoral vein. Heparin was administered for seven days at a subclinical dose while the patient was kept on bed rest with the lower extremities elevated. The edema subsided, but the pain persisted. She was discharged to wear elastic stockings and to take 5 mg of warfarin sodium per day.

Over the course of several weeks, increased pain developed in the distal extremity and the patient observed cyanosis and coldness of the affected foot. She was unaware of any edema. Several months postpartum, a vascular surgeon observed no edema or venostasis changes but found that the affected extremity was 3 to 4 degrees cooler than the normal leg. There was much tenderness in the distal calf and ankle area. Comparative x-ray films of the lower extremities showed diffuse atrophy of all the bones below the knee on the left. A lumbar sympathetic block produced pain relief for about 12 hours, but a second block was less effective. A lumbar sympathectomy was followed by complete and permanent relief of pain. The patient was fitted with a heavy knee-length elastic stocking, advised to sleep with her feet 15 cm (6 in) higher than her head, and to return gradually to her normal occupation.

Case 2

The patient, a 35-year-old male accountant and an enthusiastic runner, had tendinitis develop in one ankle for which an orthopedist applied a short cast. At first, the pain behind the ankle subsided, but about the tenth day after the cast was applied, he was awakened from sleep by severe calf pain. The following day, examination showed tenderness deep in the calf, a positive Homans' sign and some slight cyanosis. Ibuprofen was prescribed, along with elevation of the leg and the application of local heat. An examination two days later revealed essentially the same findings. The orthopedist discussed phlebography with the patient, but advised against it because of the alleged hazard of an invasive procedure. He sent the patient home on the same therapeutic regimen.

About two weeks later, increasing pain with upward extension into the thigh caused the patient to go to the emergency department of a local hospital. The emergency physician found considerable edema, a positive Homans' sign, dependent cyanosis, and engorgement of the superficial veins. A phlebogram showed obliteration of all the normal deep veins except the profunda femoris almost to the groin. The patient was admitted to hospital for two weeks on the usual regimen for acute deep thrombophlebitis. At the time of discharge, he was free of edema but had continued pain in his calf that in a few weeks became a severe burning whenever the leg was dependent.

Nine months after the acute deep vein thrombosis began, a vascular surgeon had the impression of reflex sympathetic dystrophy due to thrombophlebitis. A lumbar sympathetic block confirmed the diagnosis by providing several days of complete relief of the burning pain, but a second and a third block each provided progressively shorter pain relief. The patient then had a lumbar sympathectomy and left the hospital on the third postoperative day using a heavy knee-length elastic stocking and maintaining the usual program for venous insufficiency. With progressive exercise and then the resumption of running, he was able to resume his normal activity of running a mile or more without any leg discomfort.

Discussion

Acute deep thrombophlebitis is followed by organization of the venous thrombi and scar tissue formation with splitting of the contracting scar, so that much vein lumen is restored as a narrow irregular passage without functioning valves. The postphlebitic state results in venous insufficiency usually with the manifestations of edema increased by passive dependency, pigmentation in the distal extremity, fibrosis of the subcutaneous tissue, and, too often, stasis dermatitis and ulceration. Most patients with a postphlebitic state have little pain—only mild aching in the dependent position—as long as the skin is intact. Some patients are bothered by nocturnal cramping in the calf, and some become aware of venous claudication with exercise.

In only a few patients does the acute deep thrombophlebitis and the subsequent postphlebitic state produce the mechanism of sympathetic reflex dystrophy. The reasons are not understood. These patients usually show very few or none of the usual symptoms and signs of venous insufficiency.

The only common denominator in the small group of 14 cases of this report was the lack of prompt, vigorous treatment of the acute thrombophlebitis so that there was an extensive loss of venous valves. Ordinarily these patients should have much evidence of venous insufficiency rather than involvement of the sympathetic reflex pain mechanism without obvious signs of venous insufficiency.

The association of a disturbed emotional state with reflex sympathetic dystrophy is well known. That association was observed in most of the patients in this report, but the emotional disturbance in each case seemed to subside after the pain was relieved.

The only effective therapy in my experience involved an attack on the sympathetic innervation of the extremity. In some of the early cases, medical treatment with sympathetic inhibitor drugs was tried without success. More recently, some patients with other forms of reflex sympathetic dystrophy were given corticosteroids, but the response was so poor that this form of treatment was not considered in patients with the causalgic postphlebitic syndrome.

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